

TOURETTE SYNDROME

(Chronic Multiple Tic Disorder, Gilles de la Tourette Syndrome)

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Tourette Syndrome (TS) is a disorder characterized by uncontrolled movements and noises. It is commonly viewed in terms the difficulties it creates. Yet individuals with TS can succeed in life, not only in spite of their TS, but because of features of the syndrome. Mahmoud Abdul-Rauf (formerly Chris Jackson) is a leading free throw shooter in the NBA, an achievement he credits to the perfectionistic characteristic of his TS. Jim Eisenreich, a major league baseball player has won a number of awards for both athletic achievement and community service. Samuel Johnson, the British writer was known for his spontaneity, antics and quick wit, features often associated with TS.

Diagnostic Criteria

Tourette syndrome is a chronic movement disorder diagnosed by the presence of multiple motor and vocal tics. It begins between the ages of 2 and 15 years and is marked by a fluctuating, waxing and waning course. Typically, symptoms change slowly, with old symptoms disappearing and new symptoms replacing, or being added to preexisting symptoms. The tics commonly decrease during concentration or sleep, and increase with anxiety and tension. Tics can be suppressed, making it difficult at times for physicians to make the diagnosis. A child who only has tics when he takes medication, like Ritalin or Dexedrine, is not given the diagnosis of Tourette Syndrome.

Description of Tics

Tics are involuntary movements or vocalizations that are brief, sudden, frequent, unexpected, repetitive, purposeless, inappropriate, often irresistible and of variable intensity. Motor and vocal tics may be simple or complex.

- **Simple Motor Tics** are rapid, darting, "meaningless" movements such as eye-blinking, facial grimacing, head or nose-twitching, shoulder shrugging, abdominal tensing, frowning or rapid jerking of any body part.
- **Complex Motor Tics** are slower, more "purposeful" appearing movements that are nevertheless involuntary and typically irrelevant. These include hopping, clapping, touching objects, touching oneself or others, throwing, arranging, gyrating, bending, self-biting, rolling the eyes to the ceiling, holding funny expressions, sticking out the tongue, kissing, pinching, hitting, writing the same word perseveratively, or tearing paper or books. Echopraxia (repeating the actions of another) and copropraxia (obscene gestures) are additional examples of complex motor tics.
- **Simple Vocal Tics** are fast "meaningless" sounds such as whistling, sniffing, coughing, throat-clearing, barking, grunting, screeching, gurgling, hissing, sucking, and innumerable sounds such as uh-uh, ee-ee, and ah.

- **Complex Vocal Tics** are more recognizable, although inappropriate words, phrases or statements which are typically used repeatedly. Examples include “Shut-up”, “Stop that”, “I’m going to do better, right?” “Now you’ve seen it all right, oh boy”. Counting rituals or repeating a phrase until it is “just right” are additional examples. The most well known complex vocal tic is coprolalia, an outburst of obscene language. It probably occurs in about one third of individuals with Tourette syndrome, with a peak during adolescence. Outbursts of obscene language must be inappropriate to the situation and impulsive to qualify as tics. They typically have a “driven” quality as well.
- **Sensory Tics** have also been described. these can be sensations of pressure, tickle, warmth, cold or other abnormal sensations in the skin, bones, muscles or joints. Voluntary tightening or stretching of the muscles may temporarily relieve these tics, however, they are soon repeated.

Course of Tics

Tics most commonly appear during the early elementary years, although they may make their appearance at almost any time during childhood. The severity of tics typically waxes and wanes some each year. Overall increases in tic frequency or intensity may occur before the onset of puberty, with a decrease again during adolescence, achieving a stable pattern in adulthood.

Familial Pattern and Etiology

Tourette syndrome is an organic brain disorder with a strong genetic contribution. Boys are much more likely to be affected than girls. Multiple family members may have different symptoms of TS or the related disorders. The symptoms are thought to arise from differences in brain structure and brain chemistry. Although brain imaging studies in individuals with TS are usually normal, recent research has demonstrated a variety of differences in the size of several brain structures in individuals with TS. Some of the areas affected are the basal ganglia, frontal lobes, and limbic system. The neurochemicals which appear to be involved are dopamine, serotonin, and norepinephrine. Current research is focusing on the role of the immune system in symptom development. There are some recent reports that certain infections may precipitate or aggravate the symptoms of the disorder.

Related Features

- **Sensory Issues**
Children with TS often process sensory information differently. Under sensitivity to sensory input may result in the need for increased sensory stimulation, and seeking sensory experiences. Over sensitivity to sensory stimuli may result in the avoidance of activities which are overstimulating. Many of the behavior problems children with TS exhibit can be tied to seeking or avoiding sensory input. Some common sensory issues are hypersensitivity to certain kinds of sound (noisy areas) and/or touch (people, clothing tags), seeking of pressure (pushing, throwing, wrestling), or maintaining symmetry or balance (getting socks even, touching both sides of the body in a mirror fashion). A child who exhibits symptoms of atypical sensory functioning, or who has unexplained behavior problems, should have an assessment of sensory responses, and appropriate accommodations and interventions.

Comment [COMMENT1]: Add about sensory issues, sexual issues

■ **Educational Problems**

An increased frequency of learning disabilities is seen among children with Tourette Syndrome. Specific problems with spelling and written math are common. In addition, poor comprehension of space, time, directionality and concepts related to socialization are often observed. Motor difficulties include visual motor integration, fine motor coordination and tremors. Difficulty with handwriting is a common problem, and even when handwriting is adequate, there are often difficulties producing long written assignments and performing on timed tests.

■ **Echolalia and Echopraxia**

Children with TS may echo both speech (echolalia) and behavior (echopraxia) involuntarily. Echoing of speech may consist of repeating things others say, repeating ones own words or phrases, e.g. the end of a sentence, or unusual repetitions of specific words and phrases. Echopraxia often creates additional social difficulties for a child when the mimicry is misunderstood by others.

■ **Psychiatric Problems**

While individuals with Tourette syndrome may be well-adjusted and do well in school, an increased incidence of mental health problems is seen. Many of these are neurologically based: Attention Deficit Hyperactivity Disorder (ADHD), difficulties with peer relationships, Obsessive Compulsive Disorder(OCD), mania, and depression have all been tied to the differences in brain structure, metabolism or brain chemistry found in TS. These may be aggravated by life experiences, and lead to difficulties with self esteem, peer relationships and academic functioning.

The most common psychiatric problems seen in children with Tourette syndrome are Attention Deficit Hyperactivity Disorder (ADHD) and symptoms of Obsessive Compulsive Disorder(OCD). It is common for ADHD to be the first diagnosis made during childhood, with the diagnosis of TS being made several years later when the tics appear.

ADHD is characterized by a short attention span, motor hyperactivity, distractibility and impulsivity; it is seen in 50% to 75% of children with Tourette syndrome. It is more often seen in boys and there appears to be a positive relationship between the severity of the ADHD and the severity of the Tourette syndrome. Children with TS may be more hyperactive than other children with ADHD because of the additive effects of tics, ADHD and seeking sensory balance. The treatment of ADHD in TS is complicated by the adverse effect of some medications on the severity of tics. Even when medication is effective, classroom accommodations for sensory input, activity level, and attention span are often necessary.

Obsessive-Compulsive Disorder (OCD), is characterized by the presence of obsessions, compulsive behavior, perfectionistic tendencies, excessive orderliness and ritualistic behaviors. These compulsions and rituals may be very time consuming, and can interfere with academic achievement and domestic harmony. Other related problems seen in these children include excessive fears, panic attacks, multiple phobias and severe test anxiety. A number of medications are now available for the treatment of OCD. Psychotherapy specifically targeted at OCD symptoms is also useful in some cases.

The most common behavioral complaints are of the child having a "short-temper" and "everything being a confrontation". The child's understandable frustration over his inability to control body movements, as well as ridicule by peers, teachers and parents, plays a role in the anger and irritability that often triggers behavior problems. There is, however, evidence that anger and irritability are an intrinsic part of the disorder, at least partially due to the neurochemical makeup of the individual.

An increase in sexual interest and sexual behaviors has also been reported, and is seen in a small group of children with TS. Behaviors may be verbal or physical. Children may touch themselves or others inappropriately, expose themselves, or have sexual language either as a tic or a compulsive habit. Many times the sexual behaviors are really more infantile than strictly sexual, and can be dealt with using behavior modification techniques. Compulsive sexual speech or touching may respond to medication for tics or OCD. Children who are exhibiting complex or adult sexual behaviors should be evaluated for the possibility of being a victim of sexual abuse.

Treatment

■ **General Interventions**

Everyone dealing with TS, whether individually, as a family member, or educator will benefit from education about the nature of TS and support services. Information, support, and an extensive bibliography are available through the Tourette Syndrome Association. Children need to have a growing understanding of the disorder for their own well being and to advocate for themselves with peers, teachers, and strangers. Many will benefit from specific social skills training. Children with TS may benefit from direct mental health services, either privately or through a school based mental health program. Parents may also benefit from mental health services for emotional support and problem solving. Appropriate educational interventions for the child are essential.

■ **Medication**

Medication is indicated for treatment of tics if the symptoms are severe enough to cause psychosocial problems, difficulties with peers, teachers or family, or if they adversely affect the child's development. Medication is not a cure for TS, but it can reduce many of the distressing symptoms. Medication has been the main intervention available for the management of tics, and also plays a major role in the management of ADHD and OCD. Other features accompanying TS, like learning disorders, coordination problems, mood swings, outbursts and aggression, typically benefit little from medication. Because children with TS may have symptoms of several different conditions, more than one medication is often required. Children with TS have occasionally required three or more medications to help control their symptoms. This can be done safely, but care must be taken to avoid harmful drug interactions. The following table lists most of the medications currently in use. It should be noted that the use of many of these medications is based on research in adults, and as a consequence, unexpected responses and side effects may occur when they are prescribed for children.

A more extensive medication reference can be obtained from the Tourette Syndrome

Association, S. Calif chapter, or from the TSA internet site
<http://neuro-www2.mgh.harvard.edu/tsa/tsamain.ncll>

Tourette Syndrome Association
 42-40 Bell Blvd.
 Bayside, New York 11361-
 2820
 Tel: 718 224-2999

Tourette Syndrome Association -
 Southern California Chapter
 6545 Balboa Blvd. Suite 290
 Encino, CA 91316
 Tel: 818 344-0948

The following medication chart is provided as a brief guide to some of the medications used in the management of Tourette Syndrome and the associated disorders, along with their potential benefits and possible side effects. The information is to help families and school personnel understand the desired effects and potential side effects of the medications already prescribed for their students. It is not meant to suggest medical management for any student.

Medications Used in the Management of Tourette Syndrome

Medication	Used For			Anticipated Benefits	Disadvantages
Trade Name (generic name)	A D H D	OC D	Ti cs		
<i>Short Acting Stimulants</i>					
Ritalin (methylphenidate)	T			<ul style="list-style-type: none"> - Increased attention span. - Decreased distractibility and motor restlessness. - Decreased impulsivity. - Takes effect within an hour. - May be given daily or used only on school days. 	<ul style="list-style-type: none"> - May cause or aggravate tics. - May decrease appetite. - May cause insomnia. - Can heighten emotional lability - May lead to headaches, dizziness, abdominal discomfort, skin rash. - Growth problems may occur at high dosages. - Excessive dosage may
Dexedrine (dextroamphetamine)	T				

Medication	Used For			Anticipated Benefits	Disadvantages
Trade Name (generic name)	A D H D	OC D	Ti cs		cause decreased attention and lowered academic performance.
Long Acting Stimulants					
Ritalin SR (methylphenidate SR)	T			- As for short acting stimulants - Lasts 8 to 24 hours.	- As for short acting stimulants - Cylert must be taken daily. Liver damage has been reported - May cause or aggravate tics
Cylert (pemoline)	T				
Biphetamine	T				
Dexedrine spansule	T				
Adderal	T				
Antidepressant Group					
Tofranil ¹ (imipramine)	T			- Diminish impulsive behaviors.	- ¹ Can cause heart rhythm disturbance in high dose.
Norpramin ¹ (desipramine)	T			- Decrease hyperactivity.	- May aggravate a seizure condition.
Anafranil (clomipramine)		T		- Treat depression.	- Cannot be used if there is a risk of psychosis.
Prozac (fluoxetine)	T	T		- Help with mood disturbance.	- Needs to be taken daily.
Wellbutrin (bupropion)	T			- Improvement in compulsive and perseverative behavior.	- May cause drowsiness, nausea and constipation.
Zoloft (sertraline)	T	T		- Decrease in bed wetting.	- Tremor, especially Anafranil
Paxil (paroxetine)		T			- May increase depression.
Luvox (fluvoxamine)		T			- Possible severe side effects if used in combination with Haldol - Wellbutrin may cause or aggravate tics
Antihypertensive Group					

Medication	Used For			Anticipated Benefits	Disadvantages
Trade Name (generic name)	ADHD	OC	Tics		
Catapres (clonidine) tablets and skin patch	T	T	T	- May improve ADHD and aggressive behavior. - May also improve compulsive behaviors and tics.	- May lower blood pressure. - May cause drowsiness. - Patches may be removed and can be hazardous if swallowed - Requires monitoring of blood pressure and pulse.
Tenex (guanfacine)	T		T		
MAO inhibitors					
Eldepryl (selegiline or deprenyl)	T			- May improve ADHD symptoms in cases resistant to other medications - Limited research in TS	- Many dietary and medication restrictions
Major Tranquilizers					
Haldol (haloperidol)			T	- Primary intervention for tic control	- Drowsiness
Risperdal (risperidone)		?	T	- Risperdal may also help with mood stabilization and explosive behavior	- Involuntary movements or stiffness.
Orap (pimozide)			T	- Risperdal has limited cognitive and motor side effects	- Feeling of cognitive slowing
Prolixin (fluphenazine)			T		- Possible serious drug interactions with antidepressant group, especially Anafranil and Haldol

This chart represents a sampling of medications and is by no means comprehensive. These charts were developed by Joanne Weigel, M.D., as a brief summary review of the current literature on medications used for this purpose. A more extensive listing of medications and side effects is available in the *Tourette Syndrome Medication Reference* published by the TSA. For more information, consult the individual's medical provider.